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Case Report

Cor Triatriatum Sinister. About a Case

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Abstract

Cor triatriatum is a rare congenital anomaly. Its prevalence is 0.1% among all patients with congenital heart disease. The atrium is divided by a fibromuscular membrane in two parts; a proximal and a distal chamber that communicate with each other through two or more holes with different degrees of obstruction. It is more common to find the cor triatriatum in the left atrium (Cor triatriatum sinister).

It is usually diagnosed in childhood or during adulthood, often by chance using a routine echocardiogram. The clinical manifestations in this rare disease will depend on the degree of obstruction of the membrane in the atrium, as well as the associated congenital heart diseases.

Keywords: *Cor Triatriatum; Fibromuscular Membrane; Cor Triatriatum Sinister; Junction Rhythm*

Introduction

Literally its meaning is: "heart with three courts". It was first described in 1868 by Church and years later in 1905 Boch made a more detailed description of this malformation. Since its discovery and to date is considered a rare congenital anomaly. Its prevalence is 0.1% among all patients with congenital heart disease [1,2].

In this genetic anomaly the atrium is divided by a fibromuscular membrane in two parts; a proximal chamber and a distal chamber that communicate with each other through two or more holes with different degrees of obstruction [3]. Cor triatriatum (CT) is more common in the left atrium (cor triatriatum sinister) and is usually associated with other congenital heart diseases. The interauricular communication, the Tetralogy of Fallot, the atrio-ventricular canal, the aortic coarctation and the anomalous drainage of the pulmonary veins [4].

Detras triatriatum dexter (of the right atrium) is less frequent with an incidence of 0.025% of congenital heart diseases, and the presentation of this disease alone is uncommon [5].

Several hypotheses have been proposed to try to explain the morphogenesis of this infrequent anomaly. The first one refers to a supposed bad septation of the atrium from an abnormal development of the interatrial septum. While the second and more accepted at

present, it is sustained in the theory that a bad incorporation of the common pulmonary vein inside the atrium, is responsible for this defect [6]; as a result of incomplete absorption during the embryogenic period of the common pulmonary vein [7].

It is usually diagnosed in childhood or during adulthood, often by chance using a routine echocardiogram. The clinical manifestations in this rare disease will depend on the degree of obstruction of the membrane in the atrium, as well as the associated congenital heart diseases [8,9].

Clinical Case

It is a male patient, 45 years old, with a history of apparent health; that goes to the consultation for episode of palpitations started more than 72 hours ago. In the physical examination, he presented tachycardic, arrhythmic heart rhythms without auscultation of the presence of heart murmurs. A twelve-lead electrocardiogram (ECG) was performed and the absence of P waves, irregular RR, normal electrical axis, heart rate (HR) 145 bpm compatible with atrial fibrillation (AF) with rapid ventricular response was observed.

Frequency control is carried out in the body guard with Verapamil EV (5 mg). Once the FC was under control, maintenance dose was started with Verapamil (80 mg) 1 tab c / 8 hours, as well as oral anticoagulation with Warfarin (2 mg) 2 ½ tablet at 9 p.m. and its discharge was decided.

As part of the studies, a transthoracic echocardiogram was performed, finding the presence of a membrane that divides the left atrium (LA) into two sections; one proximal and one distal (Figure 1 and 2). In addition to the presence of a hole that connects both cavities.

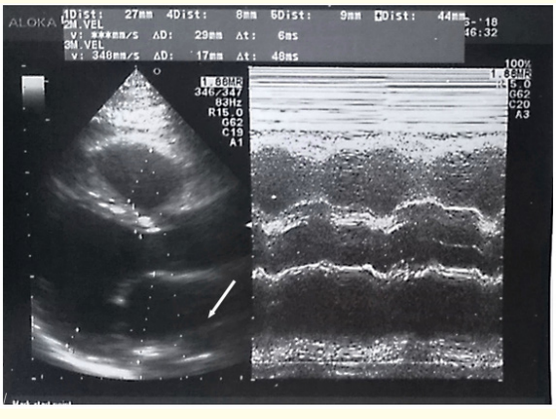

Figure 1: *Transthoracic echocardiogram, para-sternal long axis approach. (Arrow) membrane that divides the left atrium.*

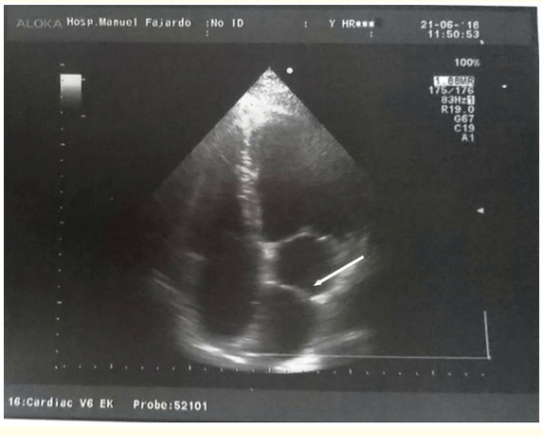

Figure 2: *Cor triatriatum sinister. Transthoracic echocardiogram, apical approach, 4-chamber view. (Arrow) Membrane that divides the AI into two chambers: proximal and distal.*

Echocardiogram

DdVD: 27 mm Ao: 29 mm ExAo: 17 mm AI: 44 mm

DdVI: 54 mm DsVI: 36 mm FEVI: 61%

Area of AI: 26 cm^2 Volume: 76 ml.

After 3 weeks of anticoagulation with therapeutic INR of 2.5 and control of FC with Verapamil (240 mg), electrical cardioversion (CVE) was performed after sedation with propofol. A 150 jule discharge is applied. The patient leaves the rhythm of the union (Figure 3) without complications, after 30 minutes of recovery in the intensive coronary care unit of the Manuel Fajardo hospital proceeds to his discharge.

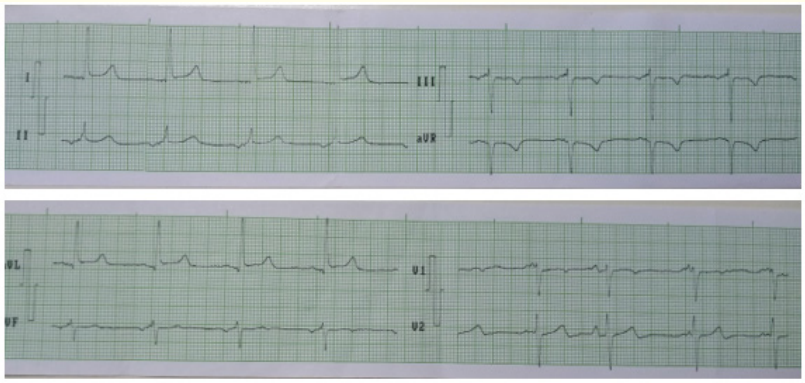

Figure 3: *Twelve-lead cardioversion electrocardiogram. Rhythm of the union.*

In the monthly follow-up consultation, the patient manifests asymptomatic, denying episodes of palpitations or dyspnea. A control electrocardiogram is performed (Figure 4) and the presence of bimodal P waves with a duration of 200 ms is confirmed, compatible with a left atrial growth, very common in patients with CT.

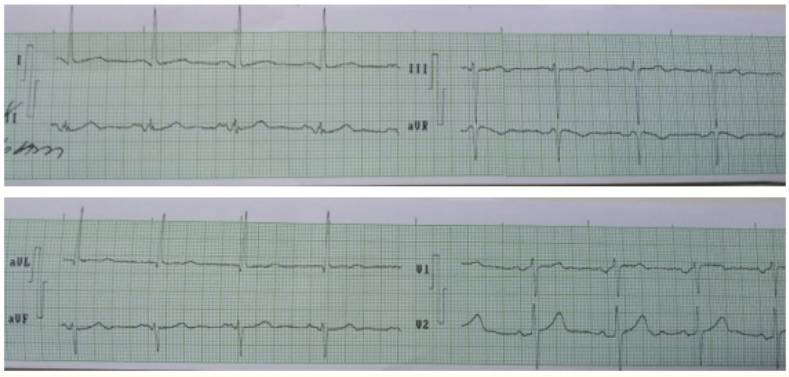

Figure 4: *Control electrocardiogram. Bi-modal P waves > 120 ms duration (left atrial growth).*

It was decided to start treatment for rhythm control with Amiodarone (200 mg); initially impregnation dose and then maintenance 1 tablet a day, in addition to maintaining oral anticoagulation with Warfarin.

Discussion

CT is an infrequent anomaly and it is uncommon for it to exceed 40 years without its surgical correction. The left atrium is divided by a fibromuscular membrane in two parts. The classic form describes the existence of a posterosuperior location chamber, where the pulmonary veins drain (upper cavity or pulmonary vein cavity). While the anterior-inferior chamber is in contact with the atrio-ventricular valve (atrial or true cavity) [10].

For the diagnosis of CT, it is essential to take into account that the atrial cavity must have the left atrial appendage and the true interatrial septum [7]. During its natural evolution, echocardiographic signs of atrial growth usually appear, as well as normal sinus rhythm disorders, a consequence of Physiological and cellular alterations of atrial tissue.

Amng the multiple classifications proposed Loeffler [11] classification is one of the most used and simple. Loeffler divides the CT into three types according to the degree of obstruction that causes the defect.

1. Type I: Not open or membrane not perforated.
2. Type II: Restrictive or fenestrated.
3. Type III: Not restrictive (only broad fenestration that communicates both cavities).

It is important to highlight that 75% of patients are diagnosed in neonatal age, mainly due to their relationship with other congenital anomalies. The natural history of this disease in patients diagnosed in adulthood is unknown [12]. They usually proceed asymptomatically until the appearance of signs or symptoms of heart failure such as dyspnea or episodes of palpitations.

In the same way that we found an AF in our patient, supraventricular tachycardias are the most frequent in CT. The origin of the tachycardia is thought to be related to the alterations in atrial anatomy that occur in this disease. These alterations affect the production and propagation of the electrical impulse from the atrial tissue.

By the beginning of the 1990s, only about 250 cases had been diagnosed worldwide. However, with the improvement of imaging techniques and the frequent use of echocardiography, the diagnosis is a little more frequent [3].

Transthoracic or tranesophageal echocardiography and the recent use of 3D Echo are the cornerstone in the diagnosis, monitoring and stratification of patients with CT [13]. However, other imaging techniques such as CT, MRI, cardiac catheterization, useful for the diagnosis and follow-up in this group of patients.

Conclusions

As usual among the cases with CT described that reach adulthood, our patient remained asymptomatic. The diagnosis is made in a fortuitous way, by going to the CG for episodes of palpitations and transthoracic echocardiography. In it, the presence of a type II CT according to the Loeffler classification with a perforated membrane without hemodynamic repercussion is confirmed.

He presented an episode of AF with rapid ventricular response that needed oral anticoagulation for three weeks, frequency control from the pharmacological treatment with Verapamil (240 mg). And in a second moment, CVE is performed to revert to sinus rhythm. After the CVE, it leaves in rhythm of the union that reverts spontaneously.

The evolution of these patients who are not surgically corrected to date is unknown. The control of the main symptoms and complications such as supraventricular tachycardias, seems until now the most viable alternative.

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